

RESIDENT ROUNDS: PART III

Primary Cutaneous Mucormycosis

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ABSTRACT

We present the case of a 36-year-old neutropenic man with acute myelogenous leukemia who presented for evaluation of a rapidly expanding necrotic eschar after adhesive placement. Histopathology revealed infection with primary cutaneous mucormycosis. Our case reviews the presentation and management of this condition as well highlights an uncommon cause in the hospital that can lead to this dangerous infection.

CASE REPORT

A 36-year-old man treated as an inpatient at an outside hospital with acute myelogenous leukemia presented to the emergency department with a 2-week history of a rapidly growing eschar on his right forearm. The lesion started at the site of adhesive tape placement for an intravenous catheter. The catheter was used only for normal saline. Once the tape was removed, the patient noticed an enlarging "black spot". The lesion was rapidly growing, slightly tender, and malodorous to the patient. He had tried no treatments.

Additional medical history included a spinal mass resulting in cord compression and leg weakness. His medications included only dexamethasone. The patient had declined all chemotherapeutic regimens in lieu of unspecified holistic treatments. Physical exam revealed a cachectic appearing male with a temperature of 100.8 degrees F and a tender 10 cm firm mass with overlying eschar on the right proximal forearm (Figure 1). Lab results revealed hemoglobin of 9 g/dL (13.8-17.2), platelets of 15,000/μL (150,000-450,000), and white blood cell count of 1.1 x 10^3/μL (3.8 – 10.8 x 10^3) with an absolute neutrophil count of 550 cells/μL. A comprehensive metabolic panel was within normal limits, including a glucose level of 105 mg/dL (<110). Punch biopsies were obtained from the border of the lesion for histopathologic analysis and culture.

Histopathologic examination of the punch biopsy revealed extensive necrotizing acute inflammation in the subcutaneous adipose tissue with broad non-septate hyphae branching at 90 degrees in the deep dermis consistent with mucormycosis (Figure 2). In addition, bacterial culture grew pseudomonas, bacteroides fragilis, staphylococcus epidermidis, and citrobacter.

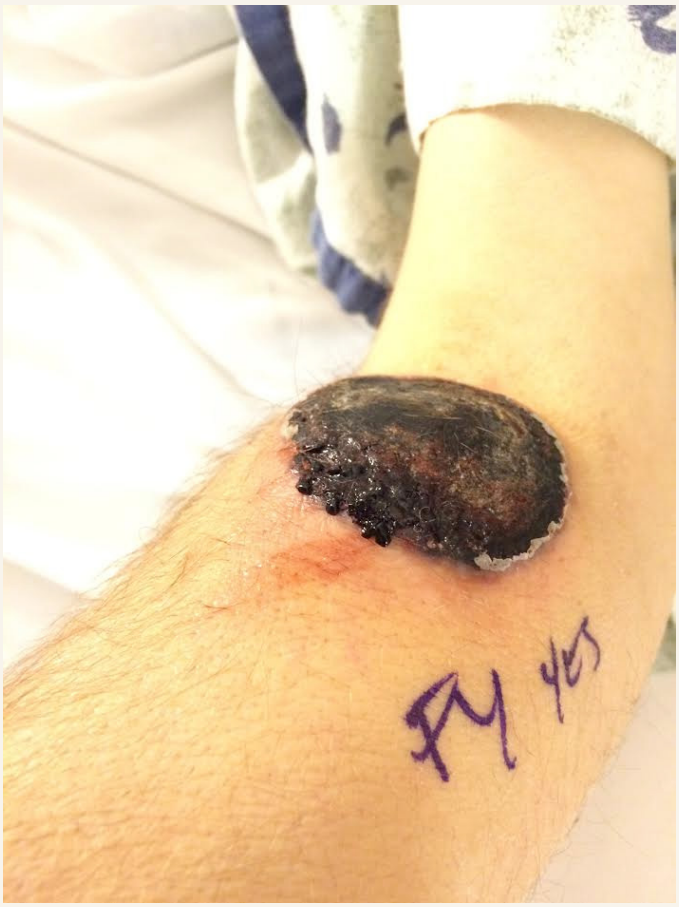
CLINICAL COURSE

Initially, the patient was treated with broad-spectrum antibiotics, including vancomycin, cefepime, and metronidazole. Once mucor was discovered via tissue biopsy, liposomal amphotericin B was started along with aggressive surgical debridements. The antibiotic regimen was narrowed to levofloxacin and metronidazole. After several debridements, no residual evidence of mucor species was clinically evident. Computed tomography (CT) scans of the head, chest, and abdomen were negative for additional abscesses. Compared to previous thoracic CT imaging, the spinal mass resolved completely. The dexamethasone was subsequently tapered to improve immune function. Repeat tissue cultures were negative. The patient was transitioned to IV posaconazole from amphotericin. When his medical condition was deemed stable, he was discharged to an acute rehabilitation unit for further medical care.

DISCUSSION

Mucormycosis, formerly known as zygomycosis, is a rare and aggressive opportunistic infection caused by saphrophytic fungi under the class Zygomycetes, order *Mucorales*, and genera *Mucor, Rhizopus, Rhizomucor, and Lichtheimia*.[1-8] This angioinvasive infection typically affects immunocompromised individuals and has a variety of manifestations including rhinocerebral (most common), pulmonary, cutaneous, gastrointestinal, and disseminated.[2,4-6,9] Primary cutaneous mucormycosis is the second most common form, accounting for

FIGURE 1. Large 10 cm mass with overlying necrotic ulceration on right forearm..

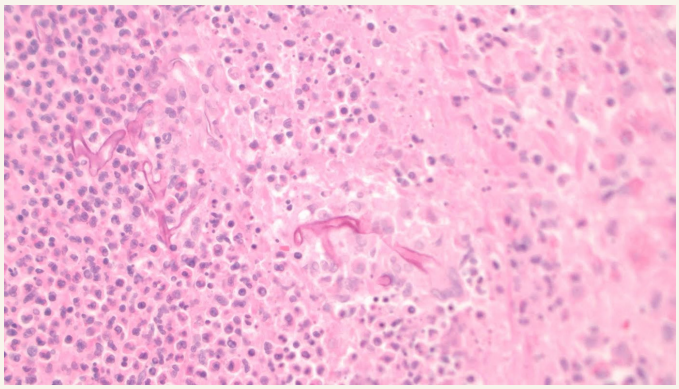


10% to 19% of mucormycosis cases, and has the best clinical outcome with a mortality rate of 15%.[2]

Mucorales species are ubiquitous in nature, found in soil and moldy foods, and contracted via inhalation of spores or direct inoculation.[1,3,6,7] Cutaneous mucormycosis may be contracted through a variety of skin wounds, including trauma, surgery, injection sites, arterial lines, insect bites, or even contaminated adhesive tapes or dressings.[2,5] Risk factors include diabetes, hematologic malignancies, allogeneic hematopoietic stem cell transplant, organ transplant, graft-versus-host disease, chronic corticosteroid use, chemotherapy or other immunosuppressive medications, deferoxamine therapy, hepatic or renal failure, intravenous drug use, and major trauma or burn wounds.[2-10]

Cutaneous mucormycosis manifests in a variety of non-specific clinical presentations, including fever, erythema, edema, vesicles, or pustules that may evolve into a necrotic ulcer, cellulitis, or gangrene.[2,5,7,8] The infection may be acute and aggressive, resembling necrotizing fasciitis, or chronic over the course of years.[2] Prompt treatment is imperative, as the infection is

FIGURE 2. Hematoxylin and eosin stain demonstrating non-septate hyphae, branching at right angles.



angioinvasive and may eventually lead to arterial necrosis, thrombosis, tissue ischemia or infarction, and dissemination.[2,4,6] Therefore, untreated infections may result in limb amputation or death.[9]

The diagnosis of mucormycosis relies on histopathology and tissue culture, as blood cultures are typically negative.[1,2,4,6] Characteristic histopathologic findings include broad (5-25 mm), non-septate hyphae, branching at 90°.[1,3,7,9] The treatment rubric involves surgical debridement, antifungals, and control of underlying conditions, such as diabetes, neutropenia, or tapering/cessation of immunosuppressant medications.[2-4, 6-9] The preferred first line antifungal therapy is liposomal amphotericin B or amphotericin B lipid complex.[1,3-5,7,8] Second-line treatment options include posaconazole5 or a combination of amphotericin B and caspofungin.[1] Recombinant growth factors may also be used in the context of neutropenic patients.[1,4] Duration of antifungal treatment is currently not well defined, but is typically conducted for at least 6 to 8 weeks.[1]

> "Primary cutaneous mucormycosis is an uncommon yet severe opportunistic infection, resulting in life-threatening angioinvasive necrotic wounds."

CONCLUSION

Primary cutaneous mucormycosis is an uncommon yet severe opportunistic infection, resulting in life-threatening angioinvasive necrotic wounds. Due to the high morbidity and mortality of this disease, prompt diagnosis, aggressive treatment, and reversal of predisposing conditions are imperative for limb salvage and survival.

DISCLOSURES

The authors have no conflicts of interest to declare.

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